What will happen when a diagnosis is made?

You will be referred to the clinic at your local Fetal Medicine Unit. You will meet with a Specialist and an ultrasound will be performed to confirm the diagnosis and note any other abnormalities. The specialist will discuss the findings with you and may recommend that you have blood tests to test for infections. You may also be offered invasive testing such as amniocentesis to test the fetal chromosomes.

The prognosis is often uncertain at best and a termination of pregnancy may be discussed with you. In some situations, further imaging with MRI scans may be recommended. Sometimes, it may be helpful to talk to another specialist such as a genetic counsellor. You will be given enough opportunity to ask questions and request further clarifications.

Our team of specialists, midwives and sonographers are always there to support you and your decisions.

For more information please contact your local NZMFMN Unit

Auckland District Health Board
Te Toka Tumai

Auckland: 09 307 4949 ext 24951

Capital & Coast District Health Board
ŪPOKO KI TE URU HAUORA

Wellington: 04 806 0774

Canterbury District Health Board
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Dandy Walker Syndrome

New Zealand Maternal Fetal Medicine Network

Reviewed: September 2015

What is Dandy-Walker Syndrome?

Dandy-Walker Syndrome is a congenital brain malformation involving the cerebellum (an area at the back of the brain that controls movement) and the fluid-filled spaces around it. The key features of this syndrome are an enlargement of the fourth ventricle (a small channel that allows fluid to flow freely between the upper and lower areas of the brain and spinal cord), a partial or complete absence of the area of the brain between the two cerebellar hemispheres (cerebellar vermis), and cyst formation near the internal base of the skull. An increase in the size of the fluid spaces surrounding the brain as well as an increase in pressure may also be present. This is known as hydrocephalus and is present in 90% of cases. It may be present post-natally.

Symptoms, which often occur in early infancy may include slow motor development. There may be signs of cerebellar dysfunction such as unsteadiness, lack of muscle coordination, or jerky movements of the eyes may occur.

Dandy-Walker Syndrome is frequently associated with disorders of other areas of the central nervous system, including absence of the area made up of nerve fibres connecting the two cerebral hemispheres (corpus callosum) and malformations of the heart, face, limbs, fingers and toes.

In about two thirds of these cases, there are other brain abnormalities. About one third of cases may have abnormalities related to other systems; the commonest association is with heart defects.

What is Dandy-Walker Complex?

Classically, posterior fossa cystic malformations have been divided into Dandy-Walker malformation, Dandy-Walker variant, mega cisterna magna, and posterior fossa arachnoid cyst. Precisely differentiating the malformations may not be possible using ultrasound or MRI. Dandy-Walker malformation, variant, and mega cisterna magna are currently believed to represent a continuum of developmental anomalies on a spectrum that has been termed the Dandy-Walker complex.

What causes this syndrome?

In most cases, the cause is not known. In some cases an autosomal recessive gene has been reported and familial occurrence has been occasionally reported. There is an association with abnormal chromosomes and a small number of cases may be associated with infections such as Toxoplasmosis and Cytomegalovirus.

Is there any treatment?

There is no treatment antenatally. Parents of children with Dandy-Walker Syndrome may benefit from genetic counselling if they intend to have more children.

What is the prognosis?

The effect of Dandy-Walker Syndrome on intellectual development is variable, with some children having normal cognition and others never achieving normal intellectual development even when the excess fluid build up is treated early and correctly.

Longevity depends on the severity of the syndrome and associated malformations. The presence of multiple congenital defects may result in severe complications or death.